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Joanna Godlewska, Justyna Orpiszewska, Wojciech Górecki

Recurrent trichobezoar in a patient with Rapunzel syndrome

Nawrotowy trichobezoar u pacjentki z zespołem Roszpunki

Department of Paediatric Surgery, Jagiellonian University Medical College, University Children Hospital, Kraków, Poland
Correspondence: Joanna Godlewska, Department of Paediatric Surgery, Jagiellonian University Medical College, Wielicka 265, 30-663 Kraków, Poland, tel.: +48 12 333 91 53, e-mail: joanna.godlewska@gmail.com

Abstract

Rapunzel syndrome is a condition where a trichobezoar is formed in the stomach and proximal intestine due to hair ingestion. A 6-year-old girl presented to emergency department with abdominal pain, vomiting and a palpable epigastric mass. Laparotomy was performed for gastric foreign body; a trichobezoar that filled the stomach, duodenum and proximal small intestine was removed. Postoperative course was uncomplicated; the patient was discharged for further out-patient follow-up and psychological care. After 7 months, the girl presented with a recurrence. A recurrent trichobezoar was removed via laparotomy. The girl was started on psychiatric treatment and iron substitution for anaemia. Ten weeks after discharge, follow-up gastroscopy was negative for gastric foreign body. There are no guidelines for follow-up after trichobezoar removal. Since the disease may be recurrent, follow-up endoscopy should be considered in order to enable an early diagnosis and less invasive treatment.

Keywords: trichobezoar, Rapunzel syndrome, gastrointestinal foreign body

Streszczenie

Zespół Roszpunki jest schorzeniem, w którym na skutek powtarzalnego spożywania włosów dochodzi do powstawania trichobezoaru wypełniającego żołądek i bliższy odcinek jelita cienkiego. Sześcioletnia dziewczynka zgłosiła się na szpitalny oddział ratunkowy z powodu bólu brzucha, wymiotów i badalnego oporu w nadbrzuszu. Ustalono rozpoznanie ciała obcego w żołądku i drogą laparotomii usunięto trichobezoar będący odlewem żołądka, dwunastnicy i bliższego odcinka jelita cienkiego. Pacjentkę wypisano w 5. dobie pooperacyjnej z zaleceniem kontroli ambulatoryjnej i opieki psychologicznej. Po 7 miesiącach dziewczynka zgłosiła się z nawrotem objawów. Wykonano laparotomię i usunięto nawrotowy trichobezoar. Włączono leczenie psychiatryczne oraz substytucję żelaza. Po 10 tygodniach od wypisu przeprowadzono kontrolną gastroskopię, w której nie stwierdzono obecności ciała obcego. Obecnie brak jest wytycznych odnośnie do postępowania pooperacyjnego po usunięciu trichobezoaru. Ze względu na możliwość nawrotowego przebiegu choroby należy rozważyć planową kontrolę endoskopową, która może pozwolić na wcześniejsze rozpoznanie nawrotu i zastosowanie mniej inwazyjnego leczenia.

Słowa kluczowe: trichobezoar, zespół Roszpunki, ciało obce w przewodzie pokarmowym

INTRODUCTION

A bezoar is a gastrointestinal foreign body resulting from deposition of indigestible matter. Bezoars are formed in the context of gastrointestinal dysmotility, altered anatomy resulting from surgical interventions, and in psychiatric conditions manifesting with eating disorders (pica). Rapunzel syndrome is a condition where repeated hair ingestion (trichophagia) leads to the formation of a trichobezoar in the stomach and duodenum, sometimes extending into the small intestine.

CASE REPORT

A previously healthy 6-year-old girl presented to our emergency department in November 2017 with abdominal pain and vomiting. On the previous day, the girl's parents noticed a protruding mass in her epigastric region. Physical examination and radiography (abdominal sonography and X-ray) yielded a diagnosis of gastric foreign body (Fig. 1). Laparotomy and gastrotomy resulted in the removal of a trichobezoar with a total length of 73 cm, filling the stomach, duodenum and proximal small intestine (Figs. 2, 3).
The postoperative course was uncomplicated; the patient was discharged on day five postoperatively. She was referred for psychological care and recommended outpatient surgical follow-up. Four months later, the girl presented again in the emergency department with abdominal pain. Physical examination, abdominal sonography and abdominal X-ray were normal; she was discharged home.

Fig. 1. Abdominal X-ray at presentation

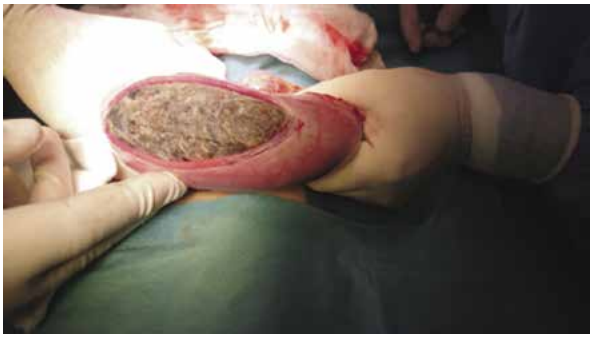

Fig. 2. Intraoperative view (first operation)

Fig. 3. Operative specimen (first operation)

After another 3 months (7 months postoperatively), the girl presented with another episode of abdominal pain and vomiting, a palpable epigastric mass and mass effect on abdominal X-ray. A recurrent trichobezoar was removed via laparotomy and gastrotomy from the stomach, duodenum and the first intestinal loop (Figs. 4, 5).
The recovery was uneventful. The consulting psychiatrist started the girl on SSRI (selective serotonin re-uptake inhibitor) treatment. Iron substitution for iron-deficiency anaemia was introduced. Social worker opinion was obtained. The patient was discharged home after 5 days with recommendation of outpatient psychiatric treatment, surgical control and gastroenterology consultation.
Ten weeks after discharge, a follow-up gastroscopy was performed, which was negative for gastric foreign body. The patient was well and asymptomatic. Pharmacotherapy is not longer required, but she continues behavioural therapy.

DISCUSSION

Rapunzel syndrome is a rare disease, most commonly affecting young girls and women suffering from psychiatric disorders[1]. Without efficient treatment of trichophagia, recurrences may occur. Only few cases of recurrent trichobezoars have been reported in the literature; they mostly concern adult patients[2–4].
The treatment of a bezoar producing acute symptoms is usually surgical via laparotomy and gastrotomy. There are reports of laparoscopic removal[5] as well as endoscopic fragmentation of a bezoar with either endoscopic removal

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Fig. 4. Intraoperative view (second operation)

Fig. 5. Operative specimen (second operation)

or leaving the fragmented mass for natural evacuation via the gastrointestinal tract[6]. However, these less invasive methods have limited application in intestinal obstruction and a large mass. In patients with no acute symptoms, there are reports of successful dissolution of bezoars with carbonated drinks such as Coca-Cola®, beer or sparkling mineral water[7], with enzymatic substances[8], with or without endoscopic assistance. Thus, elective treatment may help avoid invasive surgical procedures.

Due to the rare occurrence of trichobezoars, especially in the recurrent form, there are no widely accepted standards for patient monitoring after bezoar removal. Psychiatric and psychological care are the mainstay of treatment and are aimed at controlling trichophagia. Routine endoscopic control may permit early detection of recurrence, which in consequence could allow successful conservative management of a small mass and avoidance of repeated surgical intervention.

CONCLUSIONS

The described case is an example of an atypical, recurrent course of Rapunzel syndrome. There are no guidelines for follow-up after trichobezoar removal. Since the disease may be recurrent, follow-up endoscopy should be considered in order to enable an early diagnosis and less invasive treatment. We warn both paediatricians and family practitioners about the possibility of recurrent disease and underline the importance of continuous psychological support due to the possibility of persistent behavioural disorder.

Conflict of interest

Authors do not report any financial or personal connections with other persons or organisations, which might negatively affect the contents of this publication and/or claim authorship rights to this publication.

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